Nutcracker syndrome due to left-sided inferior vena cava compression and treated with superior mesenteric artery transposition

Bao-Zhong Yang, MD,[a] Zhen Li, MD,[b] and **Zhong-Gao Wang, MD,[b,c]** *Beijing, China*

Left renal vein hypertension secondary to left renal vein compression has been described as a cause of persistent hematuria in nutcracker syndrome. Malformation of the inferior vena cava (IVC), although rare and frequently asymptomatic, may also result in left renal vein hypertension, with resultant hematuria when it is severely compressed. We report a 20-year-old man with persistent hematuria due to compression of left-sided IVC. The patient was successfully treated by means of superior mesenteric artery (SMA) transposition and division of the fibrous bundle at the origin of the SMA. His postoperative course was uneventful. Compression of the left IVC is a unique form of nutcracker syndrome. SMA transposition, together with division of a fibrous bundle at the origin of the SMA if present, is a safe and effective surgical procedure for this special entity. (J Vasc Surg 2012;56:816-8.)

Nutcracker syndrome (NCS) commonly refers to compression of the left renal vein (LRV) between the aorta and superior mesenteric artery (SMA), resulting in increased LRV pressure, with or without hematuria. Other forms of NCS have also been reported sporadically. We successfully treated an unusual case of NCS in which LRV hypertension was caused by compression of the left-sided inferior vena cava (IVC) rather than of the LRV, as is usually found.

CASE REPORT

A 20-year-old man was hospitalized for persistent hematuria with left loin pain for 7 months. Conservative treatment proved ineffective. His symptoms worsened, and gross hematuria was present each time he urinated. Physical findings were unremarkable, except for scattered varicose veins and occasional lower limb edema after standing a long time. A urine dipstick test showed 3^+ hematuria and 2^+ proteinuria. Rare hyaline and granular cast were observed in a microscopic examination of urinary sediment.

Renal ultrasound imaging showed no abnormal findings, except that the left kidney was larger than the right kidney. A computed tomography angiography (CTA) revealed that the IVC ran upward along the left side of aorta for as long as 13 cm, received the LRV, and then crossed the aorta anteriorly through the meso-aorta angle to join the normal IVC on the right. Moreover, the meso-aorta angle was narrow (<30°), with a steep initial caudal descent. The crossing portion of the IVC was severely compressed by the SMA, causing the left-sided IVC and the LRV to be markedly dilated and the left kidney to be larger than the right kidney (Fig 1). Hematuria spraying out from the left ureter was observed on cystoscopy.

SMA transposition was performed through a left paramedian retroperitoneal approach. The spleen and descending colon were mobilized to the right, and the aorta, the SMA and its origin, the left-sided IVC, and the LRV were fully isolated (Fig 2, *A*). The SMA was transected and its distal end was reanastomosed to the infrarenal aorta in an end-to-side fashion. Extensive fibrous adhesion around the compressed portion was carefully dissected until compression of the IVC was adequately relieved. The LRV pressure reduced from 20 to 15 cm H_2O after SMA transposition and to 5 cm H_2O after thorough dissection of the extensive adhesive fibrous tissue. The tension of the left-sided IVC and the LRV was remarkably decreased (Fig 2, *B*).

The patient's postoperative course was uneventful, and the gross hematuria and microhematuria disappeared 3 and 7 days afterward, respectively. The patient was discharged 10 days after the operation without complications and resumed his studies at the university.

DISCUSSION

Malformation of the IVC, rare and often asymptomatic, has mostly been revealed through imaging examinations or has been found accidentally during abdominal aortic surgical procedures. Among various forms of IVC malformation reported, the incidence of left-sided IVC accounts for 0.2% to 0.5%.[1] It is generally accepted that the infrarenal segment derives from the right supracardinal segment. A left IVC results from the persistence of the left supracardinal vein along with regression of the right one.[2,3] Typically, the left IVC receives bilateral iliac veins, runs upward along the left side of aorta, joins the LRV at the level of the renal hila, and crosses the aorta anteriorly or posteriorly.[3,4]

From the Department of Vascular Surgery, Beijing Chao Yang Hospital of Capital University of Medical Sciences[a]; the Department of Thoracic and Cardiovascular Surgery, Second Artillery General Hospital of the People's Liberation Army[b]; and the Beijing Xuan Wu Hospital and Vascular Institute of Capital University of Medical Sciences.[c]

Author conflict of interest: none.

Correspondence: Zhong-Gao Wang, MD, FACS, Department of Thoracic and Cardiovascular Surgery, Second Artillery General Hospital of PLA, Beijing, 100088, and Beijing Xuan Wu Hospital and Vascular Institute of Capital University of Medical Sciences, Beijing, 100053, China (e-mail: zhonggaowang@yahoo.com).

The editors and reviewers of this article have no relevant financial relationships to disclose per the JVS policy that requires reviewers to decline review of any manuscript for which they may have a conflict of interest.

0741-5214/$36.00

http://dx.doi.org/10.1016/j.jvs.2012.03.021

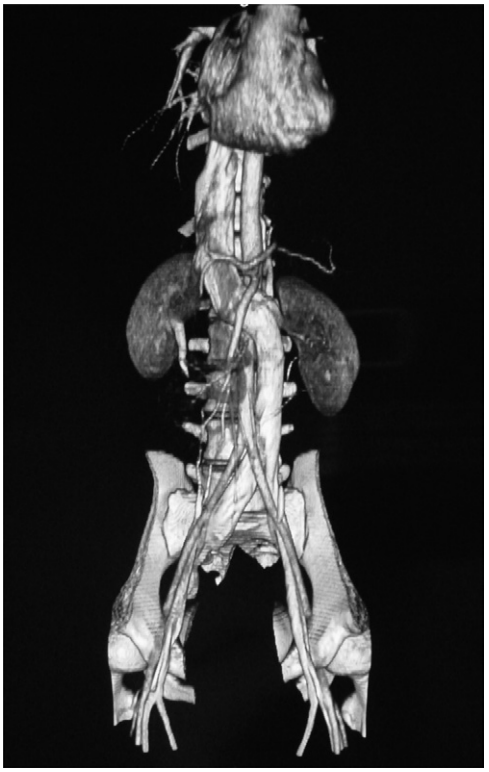

Fig 1. A volume-rendered computed tomography angiography shows compression of the left-sided inferior vena cava (IVC) by the superior mesenteric artery (SMA).

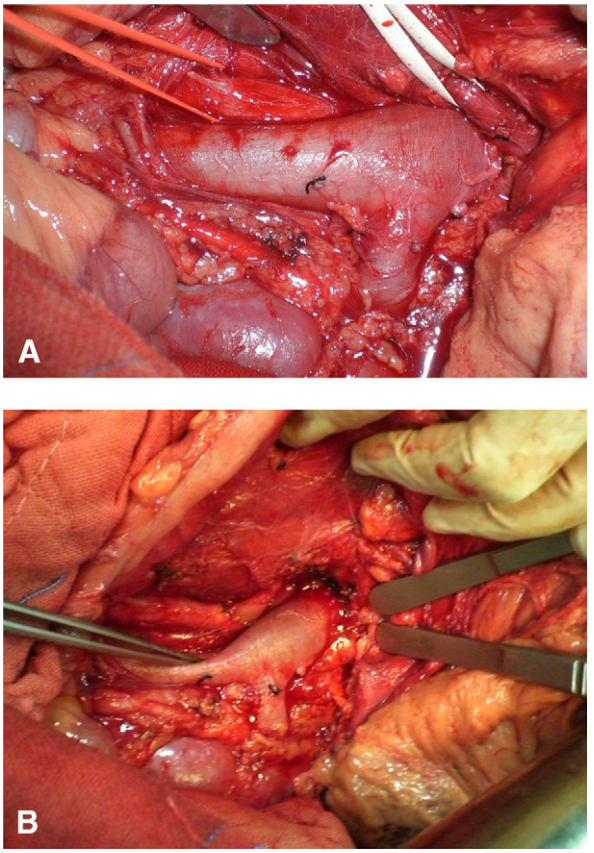

Fig 2. A, An operative photograph shows that the superior mesenteric artery (SMA) (looped with *white tape*) compressed the left-sided inferior vena cava (IVC) and created a distinctive congestion of the malformed IVC. The aorta is looped with *red tape*. **B,** The left-sided IVC was easily collapsed by a slight pressing with the tweezers tip on the left, indicating a pressure reduction. The SMA artery-to-aorta anastomosis is on the right.

Anatomically, the LRV drains into the IVC through the meso-aorta angle. The SMA normally originates from the aorta in almost a rectangular configuration so that the SMA has a 4- to 5-mm course in the ventral direction before beginning a caudal descent, thus resulting in an inverted "J" configuration. This anatomic arrangement normally prevents compression of the LRV by the SMA. An interesting finding in this patient was that the left IVC passed in front of the aorta through the meso-aorta angle, whereas the SMA branched from the aorta at an acute angle ($<30°$). These anatomic abnormalities made the IVC, rather than the LRV, severely compressed. This caused the left-sided IVC to be markedly dilated (3.07 cm) and an LRV pressure as high as 20 cm H_2O, with resultant gross hematuria and proteinuria. Excessive fibrous tissue found around the crossing portion during the operation also contributed to the compression of the IVC.

Diagnosis of NCS is difficult, often delayed, and should be established by a combination of cystoscopy, ultrasound imaging, CTA, magnetic resonance angiography, and venography. Ultrasound imaging may be used as the initial diagnostic test in patients with symptoms suggestive of NCS.[5] Although angiography and venography are accepted as the gold standard in establishing the final diagnosis of NCS, CTA, and magnetic resonance angiography, due to their noninvasiveness, have become the investigations of choice in recent years. In patients with gross hematuria, cystoscopy is indispensable in ruling out other causes and localizing the source of hematuria to the left ureteric orifice.

A number of operative and, more recently, endovascular procedures have been described to treat the typical NCS, aiming at lowering the LRV pressure by eliminating the compression of the LRV by SMA. The most commonly used procedures include transluminal balloon angioplasty and stenting of the LRV, SMA transposition, LRV transposition, and renal autotransplantation, with the latter two being the most frequently used surgical techniques. LRV transposition and renal autotransplantation were not applicable in this young patient because the site of compression was on the IVC instead of the LRV. Clearly, decompression of the IVC is the key to success in this special setting.

To our knowledge, no surgical or endovascular treatment of NCS due to left-sided IVC compression has been reported. Considering that the site of compression was on the IVC instead of the LRV in this young patient, we chose

open surgery rather than endovascular stenting because of (1) poor availability of appropriate stent at the time of surgery; (2) the long-term fate of a stent in this young patient was unclear, and (3) the geometry that would be created by a large stent in this severely compressed position was unpredictable, with potential complications of incomplete dilatation, stent collapsing, migration, thrombosis, erosion, and, possibly, rupture of the thin-walled IVC or compression of the SMA, left or right renal vein, or left IVC. In addition, we determined that a direct procedure on the IVC (replacement or bypass, for instance) would likely be more traumatic than SMA transposition.

As an already established and standardized vascular surgical procedure and valuable treatment option in patients with chronic visceral ischemia, SMA transposition, in this case, removed the "clamp" from the left IVC and permanently eliminated the "nutcracker" anatomy, without disruption of IVC continuity. This technique requires arterial clamping and reconstruction, which can be performed within a short time, thus avoiding the risk of bowel ischemia with the use of intraoperative heparin. Aspirin and antiplatelet agent (clopidogrel) were administered to prevent postoperative thrombosis.

CONCLUSIONS

The term "nutcracker syndrome" can also be used for compression of the left IVC. SMA transposition, together with division of fibrous bundle at the origin of the SMA if present, is a safe and effective surgical procedure for NCS caused by compression of the left-sided IVC. With the development of endovascular technique, use of a nitinol stent with an appropriate diameter and length, with apposition and a wide cell surface, would likely be the recommended treatment.

REFERENCES

1. Rispoli P, Conforti M, Cassatella R, Varetto G, Melloni CD, Raso AM. Left-sided inferior vena cava in patients submitted to aorto iliac surgery. Our experience and review of the literature. J Cardiovasc Surg 2001;42: 249-55.
2. Bass JE, Redwine MD, Kramer LA, Harris JH Jr. Absence of the infrarenal inferior vena cava with preservation of the suprarenal segment as revealed by CT and MR venography. AJR Am J Roentgenol 1999; 172:1610-2.
3. Bass JE, Redwine MD, Kramer LA, Huynh PT, Harris JH Jr. Spectrum of congenital anomalies of the inferior vena cava: cross-sectional imaging findings. Radiographics 2000;20:639-52.
4. Minniti S, Visentini S, Procacci C. Congenital anomalies of the venae cavae: Embryological origin, imaging features and report of three new variants. Eur Radiol 2002;12:2040-55.
5. Ahmed K, Sampath R, Khan MS. Current trends in the diagnosis and management of renal nutcracker syndrome: a review. Eur J Vasc Endovasc Surg 2006;31:410-6.

Submitted Jan 1, 2012; accepted Mar 6, 2012.